

Supplementary table S1. Comparison of characteristics of patients with primary Sjögren's syndrome (pSS) according to the 2002 American-European Consensus Group (AECG) criteria (lymphoma as exclusion criterion not employed) (n=55) and pSS patients not formally fulfilling the AECG criteria (n=50). The proportion of missing data for items in the AECG criteria was higher in those not fulfilling the criteria.

	pSS with lymphoma fulfilling 2002 AECG criteria n=55	pSS with lymphoma not fulfilling 2002 AECG criteria n=50	P value
Demographics			
Female, n (%)	48 (87)	41 (82)	0.6
Age at first reported sicca symptoms, median (range), yrs	44.5 (14-76)	50 (15-78)	ns
Age at SS diagnosis, median (range), yrs	56 (14-80)	55 (16-80)	ns
SS diagnosis in 2002 or later on, n (%)	7 (13)	5 (10)	0.8
Age at lymphoma diagnosis, median (range), yrs	64 (22-83)	64 (21-85)	ns
Overall median survival after lymphoma diagnosis, yrs (range)	8 (8 days - 20 years)	7 (0 days - 20 years)	ns
Follow-up after SS diagnosis until death or 30 October, 2013 (person yrs/ median yrs (range))	827/13 (1 year - 37 yrs)	769/14 (2 months - 39 yrs)	ns
Followed-up by rheumatologist, n (%)	44 (80)	19 (38)	0.0001
Dead at the end of the follow-up, n (%)	29 (53)	32 (64)	0.14
Clinical manifestations			
	n/available (%)		
Salivary gland swelling	34/54 (63)	22/48 (46)	0.1
Enlarged lymph nodes	18/54 (33)	16/48 (33)	1
Extraglandular manifestations*:	29/54 (54)	17/48 (35)	0.08
Arthritis	12/54 (22)	8/48 (4)	0.6
Skin vasculitis	9/54 (17)	2/48 (4)	0.06
Interstitial lung disease	3/54 (5.5)	3/48 (6)	1
Peripheral neuropathy	3/54 (5.5)	3/48 (6)	1
Myositis	5/54 (9)	0/48 (0)	0.06
Nephritis	1/54 (2)	1/48 (2)	1
Laboratory results			
ANA positivity	43/52 (83)	17/30 (57)	0.02
SSA/SSB positivity	41/52 (79)	11/22 (50)	0.02
Rheumatoid factor positivity	29/45 (64)	20/28 (71)	0.6
Anemia (haemoglobin <120g/L)	15/52 (29)	16/36 (44)	0.2
Leukopenia (leucocytes <4.0 x10 ⁹ /L)	17/51 (33)	5/36 (14)	0.05
Trombocytopenia (platelets <150 x10 ⁹ /L)	9/51 (18)	3/38 (8)	0.2
Hypergammaglobulinemia (IgG>15g/L)	28/45 (62)	13/28 (46)	0.2
Low C3 or C4	12/29 (41)	3/10 (30)	0.7
Presence of cryoglobulinemia	2/7 (29)	1/4 (25)	1
Presence of clonal component in serum	7/44 (16)	5/30 (17)	1
Positive minor salivary gland biopsy (focus score ≥1)	28/33 (85)	4/5 (80)	1
ESSDAI at SS diagnosis, median (range)**	3.5 (0-24)	4 (0-22)	ns
Immune modulating treatment for pSS***			
Any treatment given	25/54 (46)	13/49 (27)	0.04
Prednisolone ≥4 weeks	20/54 (37)	11/49 (22)	0.1
Antimalarial drugs	12/54 (22)	4/49 (8)	0.06

Sjögren's syndrome and lymphoma

Azathioprine	6/54 (11)	2/49 (4)	0.3
Methotrexate	7/54 (13)	0/49 (0)	0.01
Cyclophosphamide	3/54 (6)	2/49 (4)	1
MALT lymphoma in salivary glands, n (%)	14 (25.5)	9 (18)	0.15

* some patients with >1 manifestation; **ESSDAI= The European Sjögren's syndrome disease activity index; *** drugs below used by ≥ 5 patients during the course of pSS